CYSTS OF THE COMMON BILE DUCT.

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The following case was, clinically and pathologically, of unusual interest: P. R., a school girl, aged eight years, was sent to the University Hospital on account of persistent jaundice and an abdominal tumor. She was admitted to the service of Dr. Charles H. Frazier, to whom I am indebted for permission to use the clinical data. The following notes are abstracted from the patient's history on admission to the hospital:

Family History. Father and mother are both in good health. Three other children are living and well. One child died of cholera infantum. Father had a sore on his penis sixteen years ago. His physician cauterized it and gave him medicine during the ensuing year. There were no subsequent manifestations, and he has been well ever since. Mother had one miscarriage, about four months ago. The patient's maternal grandfather and paternal grandmother died of carcinoma of the stomach.

Social and Previous Medical History. The patient has been going to school for the past three years, and until about one year ago was always bright, strong, and active. She has had none of the acute infections of childhood, and has always been considered a healthy child.

History of Present Illness. About a year ago the patient began to be jaundiced. This jaundice was unaccompanied by pain or other manifestations of illness, except for occasional attacks of nausea. The jaundice has been persistent and has gradually progressed in severity. With the increase in the jaundice the patient's appetite has deteriorated, and there has been a moderate loss in weight. For the past eight months the stools have been clay-colored and the skin itchy. Three weeks ago the mother noted a large mass in the middle and right upper quadrants of the abdomen. The mother is sure that she would have noticed it had it been present before then. She can give no information as to any increase in size since it was first noticed. The child complains of no pain or other discomfort, and sleeps well. There are no symptoms referable to the cardiovascular, nervous, respiratory, or genito-urinary systems.

Physical Examination. The patient is a well-formed, rather poorly nourished girl of normal size for her years. She has a bright, intelligent expression. The skin of the entire body, the visible mucous membranes, and the conjunctivæ have an intense, bright icteric hue. There are no superficial glandular enlargements.

The pupils are equal and react promptly to light and accommodation. The tongue is clean and moist, and the teeth are in good condition. The examination of the heart and lungs reveals no abnormalities. The upper border of liver dulness commences at the upper border of the fourth rib in the right midclavicular line. The sharp edge of the liver can be distinctly palpated about one-half inch below the costal margin. Commencing immediately below the edge of the liver is a protuberant mass extending from the right midaxillary line to one and one-half inches below and one and one-half inches to the left of the umbilicus. It is flat to percussion, is not tender, and moves downward with inspiration. It gives the impression of being cystic. There is tympany to the right, below, and to the left of the mass. Inflation of the colon shows that organ to be situated below the mass. The palpating finger is easily inserted between the tumor and the edge of the liver. Pressure has exerted such influence on the lower right portion of the thorax as to produce marked shortening and elevation of the margin of the ribs on that side. This can be seen in the accompanying photograph (Fig. 1), in which the margin of the ribs and the boundaries of the tumor have been outlined. The spleen is not palpable and no alteration in the size or position of the kidneys can be determined.

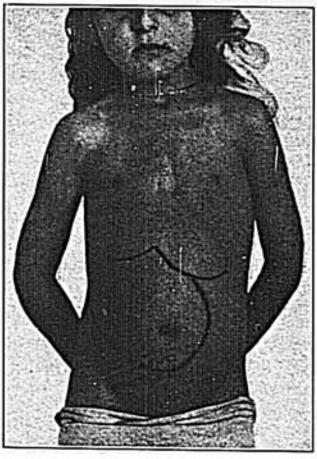

FIG. 1.—Showing the margin of the ribs and the boundaries of the tumor.

Urinalysis. Deep amber; light flocculent precipitate; acid; specific gravity, 1030; albumin and sugar negative; bile pigments positive.

Blood. Hemoglobin, 79 per cent.; erythrocytes, 4,190,000; leukocytes, 11,870. Differential count: Polymorphonuclear neutrophiles, 60 per cent.; lymphocytes, 23 per cent.; large mononuclear leukocytes, 15 per cent., transitional cells, 2 per cent.

Feces. Clay-colored; solid; faintly acid; normal odor; small number of leukocytes and epithelial cells; moderate amount of casein; occult blood negative; faint trace of urobilin.

Two days after admission to the hospital the patient was operated on by Dr. Frazier. When the mass was exposed it was found to be a cyst about the size of an ostrich egg. Coursing over its ventral surface was the stretched and flattened duodenum. The cecum and appendix were adherent to the lower pole of the cyst. The first impression was that the tumor was a cyst of the gall-bladder, but on further exploration the slightly distended gall-bladder was found lying between the cyst and the lower surface of the liver. A cannula was introduced into the cyst and about a quart of fluid resembling bile drawn off. It was then thought that the condition might be a cyst of the common duct—a view that was soon confirmed by further exploration. The adhesions were freed, the pedicle tied, and the sac removed. A dressed tube was fastened in the gall-bladder and a Mikulicz drain inserted into the wound alongside of it. In spite of vigorous stimulation, the patient failed to rally after the operation, and died three days later.

Autopsy. The autopsy, limited to the parts involved in the operation, confirmed the fact that the tumor was a cyst of the common duct. It measured 15 x 8 x 7 cm. (Fig. 2). The wall averaged about 2 mm. in thickness. The internal surface presented a peculiarly mottled appearance, by reason of the distribution of smaller and larger, irregularly shaped, brownish-green areas on a yellowish-white background. Microscopically the wall was seen to be composed of dense fibrous tissue, with an external peritoneal covering. The dark areas on the inner surface showed themselves to be more richly vascularized than the surrounding areas, and to contain deposits of bile pigments. The gall-bladder was slightly dilated; it was prevented from participating in the dilatation of the common duct by the partial obliteration of the cystic duct, resulting from compression by the cyst. The hepatic ducts were also slightly dilated. The portion of the common duct between the cyst and the duodenal papila presented itself as a firm cord-like structure, the size of a goose quill in diameter. The extramural portion measured 1 cm. in length, and was patulous; the intramural portion, which measured 2 cm. in length, was entirely devoid of a lumen. Microscopic examination of the latter portion revealed extensive fibrosis, with complete obliteration of the lumen and disappearance of all traces of epithelial elements. No opening could be found from the duodenal papilla into either the common bile duct or the duct of Wirsung, and as far as could be determined the latter suffered the same obliteration as the duodenal portion of the common duct. This view was substantiated by the histological examination of the pancreas, which showed advanced chronic perilobular pancreatitis. The liver was large, hard; and of a dark-green color. The surface was coarsely granular. Microscopically, changes characteristic of a biliary cirrhosis were seen: marked perilobular fibrosis, moderate intralobular fibrosis, dilatation of some of the intrahepatic bile ducts, and great proliferation of bile canaliculi.

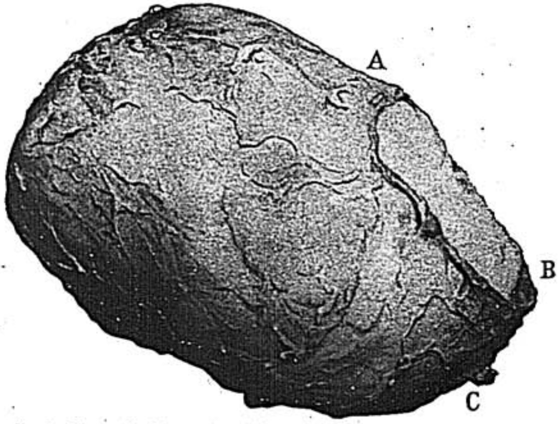

FIG. 2.—Photograph of the cyst (two-thirds natural size). *A–B*, the opening of the cystic and hepatic ducts into the cyst; *B–C*, the left wall of the common duct, forming the left wall of the cyst; *C*, the peripheral end of the common duct.

I have collected twenty-eight cases of retention cysts of the common bile duct from the literature. Some of the reports are so meagre as to make them of interest only in so far as to put the case concerned on record. Others are more elaborate, and, with the case herein reported, permit the compilation of some interesting data bearing upon the condition.

Of the 19 instances in which the sex is mentioned, 17 occurred in females. Such a preponderance of one sex is indeed striking; and yet an analysis of the cases throws no light upon the significance of this relative frequency of the condition in females. The features which tend toward the greater frequency of other diseases of the biliary passages, especially gallstones, in females are certainly not concerned in the production of cysts of the bile ducts, for these features are, for the most part, dependent upon modes of dress or functions peculiar to the female sex, possible only in adult life; and the majority of cases of cysts have occurred in the young. Of the 22 cases in which the age was mentioned, the average was fifteen years and eight months; 2 were under one year of age; 7 were between one and ten years of age; 6 between ten and twenty years of age; 4 between twenty and thirty; 1 between thirty and forty; and 2 between forty and fifty years.

Turning our attention to the anatomical peculiarities presented by the cases that have been studied, especially in regard to the lower end of the common duct and any condition there existing to cause obstruction, we find that of the 19 cases in which the result of this examination was mentioned, gallstones were found in 3, a papilloma in 1, myomyxomatous polyps in 1, scirrhous pancreatitis in 2, catarrhal cholangitis in 1, a simple stenosis or obliteration of the peripheral end of the lumen in 6, and a valve-like fold or angular insertion of the duodenal end of the duct in 5 cases. In addition, 2 of the cases in which the peripheral end of the duct was obliterated showed also this angular insertion. This great variety in the apparent cause of the obstruction and subsequent dilatation at once suggests that there is no unity of the pathogenesis of cysts of the bile ducts, but that any obstruction of the lower end of the duct may lead to cyst formation. However, let us conduct a further analysis. Of the 4 cases over twenty-one years of age in which the cause of the obstruction was mentioned, gallstones were found in 2, a papilloma in 1, and a valve-like fold in 1. Whereas, in the 13 cases under twenty-one years of age, a simple obliteration of the lumen was found in 6 instances, a valve-like fold in 4, catarrhal cholangitis in 1, and schirrhous pancreatitis in 2. I doubt the value of the opinion that scirrhous pancreatitis was the cause of the obstruction in the 2 cases so reported. They were both observed many years ago—one in 1723, the other in 1817. Considering the degree of firmness presented by the normal pancreas to palpation, and realizing the infrequency of scirrhous pancreatitis in childhood, I am loath to accept this diagnosis unsupported by a histological examination.

In considering the cases in which an obliteration of the lower end of the duct was found, we can but conjecture what the cause of this obliteration may have been. The fact that practically all these cases occurred in early life might suggest that a congenital atresia of the bile ducts was at the basis of the condition, but this is extremely improbable, for the vast majority of infants suffering with congenital atresia of the bile ducts die before the eighth month of life.[1] The fact that the case reported by Rolleston showed many manifestations of congenital syphilis, and the probable history of syphilis in the father of the patient whose case I am reporting, naturally direct attention to the possibility of the obliteration being the result of a syphilitic fibrosis. However, in the other cases there is not sufficient basis for entertaining this opinion; and it must be considered as but a vague possibility. Another possibility is that the obliteration was the result of the organization consequent upon a catarrhal cholangitis. This hypothesis seems plausible in my own case, at least, by reason of the fact that symptoms of catarrhal cholangitis preceded for some time the development of the tumor; though, of course, it must be admitted that the latter may have been present a considerable period before it was recognized by the parents. Why the duct should undergo such extensive dilatation in the presence of an obstruction in early life, as compared with the infrequency of the same occurrence in the obstructions of later life, is difficult to explain. Probably the force with which the bile is excreted is not much less in early life than in adult life, but the more delicate walls of the bile ducts of the young are less able to withstand the strain of increased internal pressure. There must, however, be some element in addition to this; otherwise it would be rational to expect to find cysts in at least some of the cases of congenital obliteration of the bile ducts in which the obstruction is in the lower end of the duct; I am not aware of any such case having been reported. The other element in the production of the cysts is, in all probability, a congenital weakness of the muscular tissue of the wall of the duct.

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[1] Lavenson, Jour. Med. Research, 1908, civ, 61.

In the cases of Douglas, Arnolds, Fenger, Arnison, Rostowzew, and Konitzky, and in my case, the lower end of the duct, or its fibrous remains, was found to enter the cyst at so acute an angle as to produce a valve-like formation at the duodenal end of the cyst. Rostowzew looks upon this angular insertion of the duct as the cause of the obstruction and the subsequent dilatation. His view can be best appreciated by glancing at the accompanying diagrams. As is shown in Fig. 3, the intramural portion of the common duct runs normally from left to right; in other words, in the same direction as the portion of the duct above the duodenum, but at a somewhat more acute angle. Rostowzew believes that in some cases the intramural portion of the duct, instead of running from left to right, runs from right to left, as is shown in Fig. 4. This according to his view produces a kink, which results in obstruction to the outflow of bile and subsequent cyst formation, as is shown in Fig. 5. I believe Rostowzew's hypothesis to be incorrect. In my opinion the angular insertion of the duct is the result of the cyst formation, and not its cause. If one recalls the anatomical relations of the common bile duct as it runs in the gastrohepatic omentum, it will be appreciated that lying farthermost to the right as it does, any tendency to dilatation that it may assume toward the left would have to overcome the resistance offered by the hepatic artery and the portal vein; whereas, to the right no structures are interposed to restrict its dilatation.

In my case, in which, in addition to obliteration of the duodenal end of the duct, this angular insertion was found, it can be seen by glancing at the accompanying photograph of the cyst (Fig. 2) that the dilatation was entirely to the right. The short straight line *B*-*C* represents the left wall of the common duct, the right wall alone being distended to form the cyst. With such extensive dilatation to the right, the peripheral undilated portion of the duct is compelled to change its direction, and instead of running from left to right, as it normally does, to run sharply from right to left, as is shown in Fig. 6.

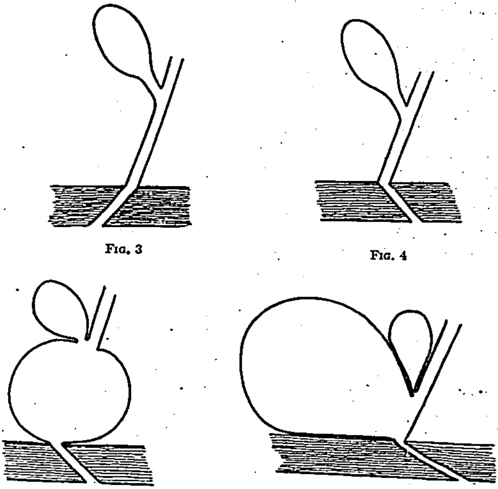

FIG. 3

FIG. 4

FIG. 5

FIG. 6

FIGS. 3, 4, 5, and 6.—The varying modes of insertion of the common bile duct into the duodenum.

This view is substantiated by the fact that in 3 of the 7 cases in which this angular insertion was present, obliteration or obstruction of the peripheral portion of the lumen was also found. In the other 4 cases it is not specifically mentioned whether or not the peripheral portion of the duct was patulous. Assuming that it was patulous, I believe it probable that in the presence of a congenital weakness of the muscular tissues of the duct wall a temporary obstruction, as by a catarrhal cholangitis or a mucous plug, could offer sufficient resistance to the outflow of bile to result in dilatation and a consequent angular insertion of the lower end of the duct. Even though the cause of the obstruction should subsequently subside, by the time the cyst is formed the valve-like opening could undoubtedly act as a secondary cause of obstruction.

SYMPTOMATOLOGY. There is little to be said of the symptomatology of the condition, aside from what has already been mentioned in discussing its pathology and pathogenesis. It should be recalled that in many of the cases symptoms of biliary obstruction have preceded the formation of a cyst, or, probably better, the recognition of a cyst, by a considerable period of time. Recognition must also be taken of the fact that it occurs relatively much more frequently in childhood and in the female sex. The conditions with which it is most likely to be confused are cysts of the pancreas and dilatation of the gall-bladder. Probably, in the vast majority of cases, only an exploratory incision will determine the diagnosis.

TREATMENT. It can be readily appreciated that surgery offers the only possibility of cure. Of the 21 cases in which more or less extensive surgical procedures were adopted, simple puncture of the cyst was performed in 3 cases, all of which resulted in death. Incision and drainage was performed in 14 cases, with 13 deaths. Cholecystenterostomy was performed in 4 cases, with 3 recoveries. The latter operation thus seems to offer the best chances of cure.

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